Fenestration Between the Left Atrial Appendage and Left Superior Vena Cava in Kawashima Operation

Rıza Turkoz, MD, Oğuz Omay, MD, Canan Ayabakan, MD, Can Vuran, MD, Kursat Tokel, MD, İlhan Günay, MD, and Sait Aşlamaci, MD

Departments of Cardiovascular Surgery and Pediatrics, Baskent University, İstanbul Teaching and Medical Research Center, İstanbul, Turkey

The benefits of a baffle fenestration in essentially high-risk Fontan patients have been demonstrated. Described here is the use of a new fenestration between the left atrial appendage and the left superior vena cava after Kawashima operation in a patient with a double-outlet right ventricle with hypoplastic left ventricle, left atrial isomerism, bilateral superior vena cavae with no bridging vein, an interrupted inferior vena cava, and continuation of the hemiazygos vein to the left superior vena cava.


© 2008 by The Society of Thoracic Surgeons

High-risk Fontan candidates who have the staged Fontan procedure (first bi-directional cavopulmonary anastomosis and then total cavopulmonary connection) have excellent clinical results [1]. However, patients with single ventricle, left atrial isomerism, interruption of the inferior vena cava, and bilateral superior vena cavae are often palliated with Kawashima operation. This operation consists of bilateral cavopulmonary anastomoses in which hepatic venous flow is left within the systemic circulation. Patients with a mean pulmonary arterial pressure of more than 20 mm Hg are challenging because of the risk of cavopulmonary dysfunction. The benefits of a baffle fenestration in essentially high-risk Fontan patients have been shown [2]. However, creation of a classic baffle fenestration is not technically possible during the Kawashima operation. We present the unique case of a patient with borderline mean pulmonary artery pressure who underwent a Kawashima operation and fenestration between the left atrial appendage and the left superior vena cava.

Technique
A cyanotic newborn was diagnosed as having left atrial isomerism, a double-outlet right ventricle, a hypoplastic left ventricle, a large ventricular septal defect, an atrial septal defect, and pulmonary stenosis. When he was 1.5 months old, he received palliative treatment at another institution with a left modified Blalock-Taussig shunt. He was 6 years old when he presented to our clinic. Cardiac catheterization confirmed the diagnosis showing bilateral superior vena cavae with no bridging vein, an interrupted inferior vena cava, and continuation of the hemiazygos vein to the left superior vena cava. The systolic blood pressure in the right ventricle was 123 mm Hg, and it was 104 mm Hg in the aorta. The mean pulmonary artery blood pressure was 22 mm Hg.

A Kawashima operation (bilateral cavopulmonary anastomosis) was planned, but because the mean pulmonary artery pressure was high, the operation was considered to be high risk. After standard aortic cannulation and venous cannulation through both superior vena cavae and the right atrium, cardiopulmonary bypass was begun. The main pulmonary artery and the modified Blalock-Taussig shunt were divided. The pulmonary artery branches were dissected distally and the bilateral superior vena cavae were anastomosed to the branch pulmonary arteries. A 3-mm fenestration was made using side clamps and directly anastomosing the left atrial appendage to the left superior vena cava at its cannulation site (Fig 1). After modified ultrafiltration, the pressure measurements were 75 mm Hg systolic blood pressure in the aorta, 8 to 9 mm Hg mean blood pressure in both superior vena cavae, and 4 mm Hg mean pulmonary blood pressure in the right atrium. The transpulmonary gradient was calculated as 5 mm Hg. The oxygen saturation postoperatively ranged from 80% to 86%. The bilateral chest drains were removed on day 4 after surgery, and the patient was discharged on day 8 after surgery. At 7-month follow-up, the patient is in good condition with oxygen saturation of 83%.

Comment
Hannan and associates [3] previously described a fenestrated Kawashima operation in which a calibrated 3-mm connection between the right pulmonary artery and the
A common atrium was constructed with the proximal right superior vena cava. This technique leaves a short distal superior vena cava for a pulmonary artery anastomosis, therefore creating a wide bi-directional anastomosis may be difficult. In patients with an ipsilateral Blalock-Taussig shunt and right pulmonary artery stenosis, a longer distal superior vena cava is preferred and will make for better reconstruction at the pulmonary artery site.

Picarelli and associates [4] have done a Kawashima operation in which they make a fenestration between the right atrial appendage and the right pulmonary artery. This technique is easier if the right atrial appendage is wide and long; however, the right atrial appendage is usually situated anterior and inferior to the right pulmonary artery, making the anastomosis difficult, whereas the left atrial appendage is almost always near the left superior vena cava. There is greater risk of a pulmonary arteries stenosis when the fenestration is created directly on the pulmonary arteries during a Fontan operation. In our fenestration technique, after weaning from cardiopulmonary bypass, a direct connection is easily created by placing two side clamps on the left atrial appendage and the left superior vena cava with no risk of pulmonary artery distortion and stenosis.

In children with an interrupted inferior vena cava and azygous continuation, Kawashima operation eliminates the hepatic venous effluent from the pulmonary circulation. Consequently this may cause pulmonary arteriovenous malformations to develop. The incidence of pulmonary arteriovenous malformations has been reported to be as high as 58% within 5 years after the Kawashima operation [5]. Brown and colleagues [5] analyzed the development of pulmonary arteriovenous malformations in their patients after the Kawashima operation. They revealed that none of their patients who had undergone completion Fontan within 18 months had evidence of pulmonary arteriovenous malformations. Furthermore, interval between Kawashima operation and completion Fontan of more than 2 years was found as the most sensitive predictor of development of pulmonary arteriovenous malformations. Therefore, as with Brown and colleagues [5], we recommend early redirection of the hepatic venous effluent to the pulmonary arterial circulation. Because transcatheter occlusion of the fenestration between the left atrial appendage and the left superior vena cava is technically difficult, redirecting the hepatic venous effluent to the pulmonary arteries and occluding the fenestration will be performed simultaneously in a second operation within 2 years of the Kawashima operation.

Creating a fenestration between the left atrial appendage and the left superior vena cava is a novel technique in a Kawashima operation. The technique is simple, quick, and without risk of a pulmonary artery stenosis and makes a reliably effective fenestration for patients with high-risk hemodynamics.

References